Dysferlin polyclonal antibody

Catalog: BS90435 Host: Rabbit Reactivity: Human, Mouse

BackGround:

Dysferlin is a muscle-specific protein that is essential for normal muscle function and development. Mutations in the human dysferlin gene, DYSF, which maps to chromosome 2p13.2, are associated with limb girdle muscular dystrophy-2B (LGMD-2B) and a related, adult-onset, distal dystrophy known as Miyoshi myopathy (MM). Dysferlin localizes to the muscle fiber membrane, but is absent in MM and LGMD-2B muscle. Dysferlin is detected in 5-6 week embryos, when limbs begin to form regional differentiation. Although it is not essential for initial myogenesis, dysferlin appears to be critical for sustained normal function in mature muscle. It has been suggested that the absence of dysferlin during development gives rise to the disease phenotype in adulthood. Identical mutations in the dysferlin gene can produce more than one myopathy phenotype, indicating that additional genes and/or other factors are also involved in the clinical phenotype. The DYSF gene has no homology to any other known mammalian gene, but the protein product is related to the spermatogenesis factor fer-1 of Caenorhabditis elegans. The name “dysferlin” combines the role of the gene in producing muscular dystrophy with its homology to C. elegans.

Product:

Rabbit IgG, 1mg/ml in PBS with 0.02% sodium azide, 50% glycerol, pH7.2

Molecular Weight:

237 kDa

Swiss-Prot:

O75923(Human) Q9ESD7(Mouse)

Purification&Purity:

ProA affinity purified

Applications:

WB:1:1,000-1:5,000

ICC:1:50-1:200

IHC:1:50-1:200

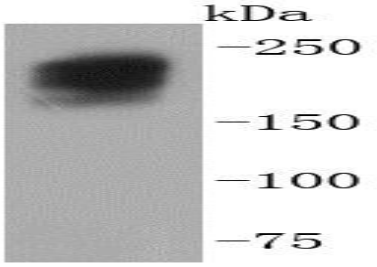
Storage&Stability:

Store at +4℃ after thawing. Aliquot store at -20℃ or -80℃. Avoid repeated freeze / thaw cycles.

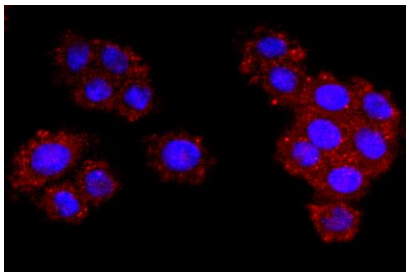
Specificity:

Dysferlin polyclonal antibody detects endogenous levels of Dysferlin protein.

DATA:



Western blot analysis of Dysferlin on human skeletal muscle lysates using anti-Dysferlin antibody at 1/1,000 dilution.



ICC staining Dysferlin in SW480 cells (red). The nuclear counter stain is DAPI (blue). Cells were fixed in paraformaldehyde, permeabilised with 0.25% Triton X100/PBS.

Note:

For research use only, not for use in diagnostic procedure.

Bioworld Technology, Inc.
Add: 1660 South Highway 100, Suite 500 St. Louis Park, MN 55416,USA.
Email: info@bioworlde.com
Tel: 6123263284
Fax: 6122933841

Bioworld technology, co. Ltd.
Add: No 9, weidi road Qixia District Nanjing, 210046, P. R. China.
Email: info@biogot.com
Tel: 0086-025-68037686
Fax: 0086-025-68035151